A woman in her 50s was admitted after several months of progressive dyspnea on exertion and severe lower extremity edema. She had undergone aortic valve replacement for aortic stenosis, 2-vessel coronary artery bypass, and permanent pacemaker placement 1 year before presentation. She had a history of mantle chest radiotherapy for Hodgkin lymphoma at age 17 years, which was curative. After successful intravenous diuresis, neck examination at approximately 60° (Figure 1A and Video) revealed a regular carotid pulsation and discernible internal jugular venous pulse. After examination of the neck at rest, the patient took a small breath and held it for a few seconds. Hemodynamic data at left and right heart catheterization are shown in Figure 1B.

**WHAT IS YOUR DIAGNOSIS?**

A. Tamponade
B. Myocardial restriction
C. Right ventricular failure
D. Wide-open tricuspid regurgitation
Diagnosis

B. Myocardial restriction

Discussion

When examining the venous pulsation (Video and Figure 1A), one notes a clear α wave just before the carotid systolic pulsation, followed by a visible x descent and a noticeable v wave coincident with the end of carotid pulsation. The absence of a large c-v wave abolishing the x descent (right atrial ventricularization) suggests that severe (wide-open) tricuspid regurgitation is not present. Immediately after the carotid systolic pulsation, there is an abrupt disappearance of the venous pulsations representing a rapid or deep y wave descent (Figure 2A). A rapid y descent is suggestive of a constrictive or restrictive process and coincides with the beginning of the “square root sign” (rapid increase in right and left ventricular diastolic pressure immediately after systole [Figure 2B, arrowhead]) during right-left heart catheterization. Conversely, tamponade is associated with a blunted y descent reflecting continuous elevation of diastolic pressures, without significant phasic variations. A prominent Kussmaul sign was then observed on inspiration (Video and Figure 2B, arrowhead), but this sign is nonspecific and could be seen in all 4 proposed diagnoses since it represents elevated right-sided pressures (irrespective of cause) that increase with inspiration as blood rushes into the right-sided chambers. The differentiation between constrictive and restrictive physiologic states cannot be entirely made by isolated neck vein observation, since both processes will likely exhibit jugular venous distention, rapid inspiration, and concomitant cytotoxic chemotherapy.2,3 As such, history of chest irradiation in a patient presenting with heart failure symptoms should prompt evaluation of coronary, myocardial, pericardial, and valvular heart disease due to tissue fibrosis. Risk factors for radiation-induced cardiotoxicity include radiation dose greater than 30 to 35 Gy, greater field size, younger age at exposure, longer time since exposure, and concomitant cytotoxic chemotherapy.4,5

Survivors of Hodgkin lymphoma are at a substantially increased risk of developing radiation-related cardiovascular complications throughout their lives.1 Radiotherapy for Hodgkin lymphoma has been associated with coronary, myocardial, pericardial, and valvular heart disease due to tissue fibrosis. Risk factors for radiation-induced cardiotoxicity include radiation dose greater than 30 to 35 Gy, greater field size, younger age at exposure, longer time since exposure, and concomitant cytotoxic chemotherapy.2,3,5,6 As such, history of chest irradiation in a patient presenting with heart failure symptoms should prompt evaluation of coronary, myocardial, pericardial, and valvular heart disease as well as pulmonary disease (our patient had moderate restrictive lung disease demonstrated by pulmonary function tests). Of these conditions, the differentiation between restrictive cardiomyopathy and constrictive pericarditis can be challenging but invaluable. Constrictive pericarditis may be curable by pericardiectomy, whereas restrictive cardiomyopathy is not and thus is managed medically.

Transesophageal echocardiography is the initial noninvasive diagnostic test of choice when evaluating patients for restrictive cardiomyopathy and/or constrictive pericarditis. Various criteria exist in differentiating constrictive pericarditis from restrictive cardiomyopathy by echocardiography.4 However, in a subset of patients, echocardiography may not be able to distinguish restrictive cardiomyopathy from constrictive pericarditis and hemodynamic catheterization may be required.5

A follow-up echocardiogram 1 year later showed increasing pulmonary hypertension, mild mitral stenosis, unchanged tricuspid regurgitation, and a normally functioning aortic valve prosthesis.

ARTICLE INFORMATION

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REFERENCES